

Renal leiomyosarcoma: A rare entity

Renal leiomyosarcoma

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Abstract

Renal leiomyosarcoma is one of the rare tumors. It constitutes only 0.5% of all primary renal malignancies. It accounts for 50-60% of primary renal sarcomas. The majority of cases are adults, and their mean age is 58 years. Renal leiomyosarcomas originate from the renal capsule, pelvis renalis, calyces, and vascular smooth muscles. These tumors have a poor prognosis, and surgery is the main treatment modality. In this article, in the light of the literature, we aimed to present a case report of renal leiomyosarcoma 16x15x11 cm in size in the right kidney of a 39-year-old female patient who presented with complaints of abdominal pain and hematuria. Although leiomyosarcomas are limited in the kidney, they are aggressive tumors with a poor prognosis.

Keywords

Leiomyosarcoma; Kidney; Aggressive tumors

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Introduction

Sarcomas are rarely encountered tumors, and renal sarcomas observed in adults include leiomyosarcoma, osteosarcoma, angiosarcoma, rhabdomyosarcoma, synovial sarcoma, pleomorphic sarcoma, and fibrosarcoma.

Primary leiomyosarcomas are rare tumors observed in adults, and these tumors, which show smooth muscle differentiation, are the most common renal sarcomas. Primary sarcomas in the kidney constitute 0.8-2.7% of renal tumors in adults. Leiomyosarcomas are aggressive and highly malignant tumors, and they originate from the renal capsule, pelvis renalis, calyces, and vascular smooth muscles. Although they have an equal prevalence in males and females, in some publications in the literature, a higher prevalence is reported for females. Leiomyosarcoma is observed at a younger age than classic renal cell carcinoma. It does not have any specific clinical findings, and patients most commonly present with abdominal pain, hematuria, and weight loss. The differential diagnosis includes angiomyolipoma, sarcomatoid renal cell carcinoma, and malignant melanoma. The radiological findings are nonspecific, and the diagnosis is made by a postoperative pathological examination. Leiomyosarcomas are aggressive tumors, and their 5-year survival rate is around 29-36% [1].

Case Report

When a 39-year-old female patient was admitted to the urology clinic of our hospital with complaints of abdominal pain and hematuria, the physical examination revealed a firm, well-circumscribed mass on the right side. Renal color Doppler ultrasonography revealed a solid space-occupying lesion 134x72 mm in size, with discrete contours and heterogeneous echotexture, showing discrete arterial and venous vascularization on Doppler examination, and originating from the right kidney and showing an exophytic medial extension (hypernephroma?). Magnetic resonance (MR) examination of the upper abdominal revealed parapelvic T1A hypointense and T2A hyperintense heterogeneous mass lesion with cystic cavities, approximately 162x106 mm in diameter, showing an exophytic extension and applying lateral pressure to the vena cava in the right kidney. Right radical nephrectomy was applied to the patient. The patient, who did not develop complications, was discharged on the 4th postoperative day with stable vital signs and laboratory values. The nephrectomy material which was sent to the pathology laboratory was 16x12x11cm in size and 1890g in weight. In the cross-section of the kidney, a solid cystic tumoral formation 15x12x11cm in size was observed in a gray-beige colored, well-circumscribed area based on the renal capsule. Necrotic areas were observed in serial sections. When examining multiple sections of the mass, the tumor structure was observed at a distance of 0.2 cm, when closest to the renal capsule, not exceeding the capsule, with well-circumscribed focal necrosis. It was observed that the tumor was composed of pleomorphic, large eosinophilic cytoplasmic spindle cells with a bizarre-looking nucleus in places that form intercrossing bundles (Figure 1A-B). Six-seven mitotic figures were observed in the 10 large augmentation areas (LAA) in the tumor. Capsule and perirenal fat tissue invasion was not observed. No lymphovascular invasion was observed. In the

immunohistochemical staining process that was performed for the differential diagnosis, there was positive staining in tumor cells with smooth muscle actin (SMA) (Figure 2A), vimentin, desmin (Figure B), while there was negative staining with epithelial membrane antigen (EMA), S100, CD117, CD34, myoglobin, and mouse anti-human melanoma antibody -45 (HMB45). The rate of proliferation was found to be 60% with Ki67.

The patient was diagnosed as renal leiomyosarcoma with histopathological findings and immunohistochemical staining results. There was no diagnosed sarcoma in another localization. When the tumor was graded by the French Federation of Cancer Centers Sarcoma Group (FNCLCC) (Table 1), tumor differentiation was reported as a score: 1 (sarcoma closely resembling normal adult mesenchymal tissue), tumor necrosis score: 1 (<50%), mitotic score: 1 (0-9 mitoses per 10 HPF), histological grade: 3 (total score: 3).

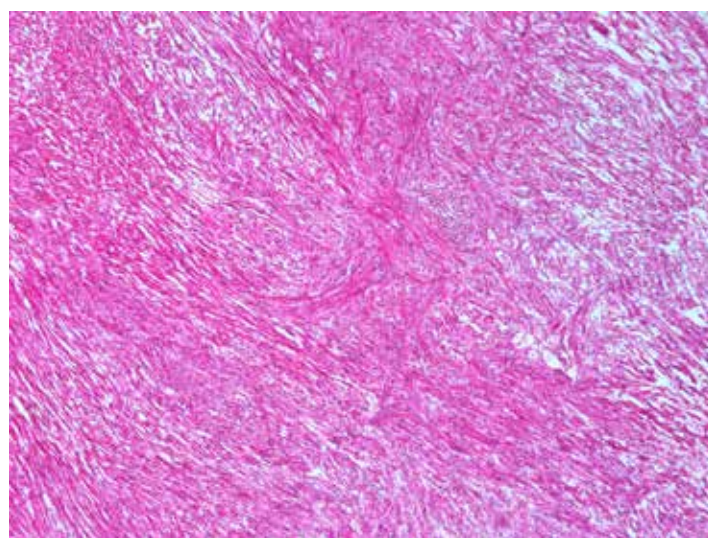


Figure 1A. Fascicular arrangement of spindled cells is seen. (H&E, 200)

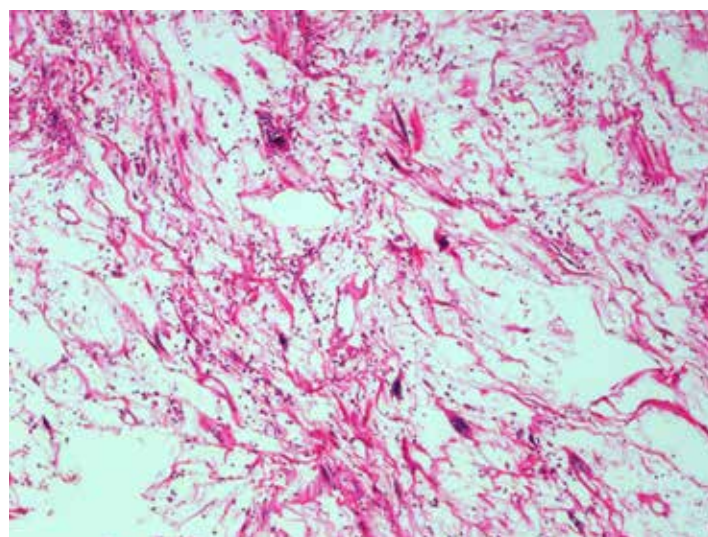


Figure 1B. Elongated/ spindle cells with cigar-shaped nuclei and a moderate amount of bright eosinophilic cytoplasm (H&E, 200).

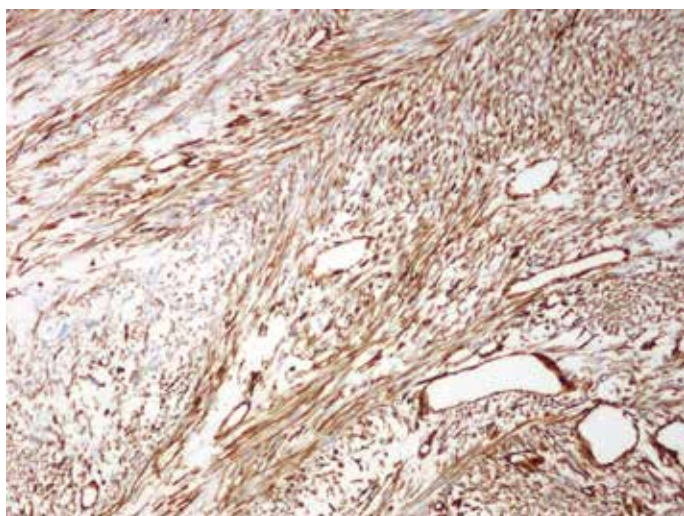


Figure 2A. Positive staining in tumoral cells with A; (SMA, x200)

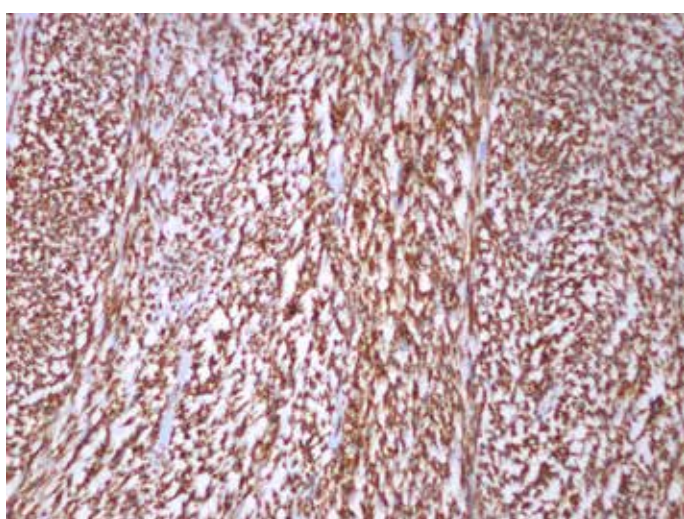


Figure 2B. Positive staining in tumoral cells with A; (Desmin, x200)

Table 1. Classification of the histological grade used by the French Federation of Cancer Centers Sarcoma Group (FNCLCC)

Tumor differentiation	
Score 1	Sarcoma closely resembling normal adult mesenchymal tissue
Score 2	Sarcoma for which histologic typing is certain
Score 3	In myxoid variants, expression may be weaker
Mitotic Count	
Score 1	0-9 mitoses/10HPF
Score 2	10-19 mitoses/10HPF
Score 3	≥ 20 mitoses/10HPF
Tumor necrosis	
Score 0	No necrosis
Score 1	<50% tumor necrosis
Score 2	>50% tumor necrosis
Histologic grade	
Score 1	Total score: 2 to 3
Score 2	Total score: 4 to 5
Score 3	Total score: 6 to 8

Discussion

Renal leiomyosarcomas are very rare tumors. Primary sarcomas in the kidneys constitute 0.8-2.7% of renal tumors in adults. These are the most common renal sarcoma subtypes (50-60%). They are mostly encountered in adults in the 5th-6th decade. While males and females are equally affected, in some publications in the literature, a higher prevalence is reported for females [2]. Leiomyosarcomas are solitary lesions, and abdominal pain, hematuria, and palpable mass are the main symptoms. The tumor is often localized on the right side [4]. The tumor originates from the renal capsule, pelvis renalis, calyces, and vascular smooth muscles. The most common origin is the smooth muscles of the renal vein [3]. It was located in the right kidney of our female patient and probably originated from the renal capsule.

The first and most common symptoms are abdominal pain and hematuria, while others are palpable mass and weight loss. In our case, hematuria and abdominal pain were the first clinical complaints.

Radiological examinations such as ultrasonography, tomography and magnetic resonance cannot differentiate leiomyosarcoma from renal cell carcinoma. The diagnosis is made by postoperative pathological examination.

Renal leiomyosarcomas are solid, gray-beige colored, well-circumscribed, necrosis-containing masses with a diameter of 5 to 17 cm. Most tumors have cystic changes [1]. Our case was 16 cm in diameter, and cystic changes and necrotic areas were observed.

Microscopy revealed fascicular, plexiform, spindle cell bundles exhibiting a random growth pattern with nuclear pleomorphism, mitosis, necrosis, and mitotic figures. In the differential diagnosis, sarcomatoid renal cell carcinoma and angiomyolipoma are important. Leiomyosarcoma is stained with desmin, vimentin, and SMA. The majority of tumors are negative for cytokeratin, CD34, CD117, EMA, and S100, although (uncommonly) focal aberrant expression of these markers can occur. Melanocytic markers (HMB45, and Melan-A), Myo-D1, and myogenin are negative. This staining profile supported a smooth muscle origin and confirmed the diagnosis of leiomyosarcoma. The tumor proliferation rate was calculated as 60% with ki67 immunohistochemical staining performed.

Renal leiomyosarcoma can be distinguished from leiomyoma and angiomyolipoma, by the presence of cellular pleomorphism, increased mitosis, and necrosis [5]. Angiomyolipomas are triphasic tumors, may have degenerative changes, but no marked atypia, no mitotic activity, positive for melanocytic markers.

High-grade tumors are pleomorphic, necrotic, requiring immunohistochemical stains to distinguish them from other tumors such as sarcomatoid carcinoma pleomorphic sarcomas, sarcomatoid renal cell carcinoma. While leiomyosarcomas contain predominantly monomorphic cells, sarcomatoid renal cell carcinomas contain pleomorphic cells and lack the typical fascicles of smooth muscle cells seen in leiomyosarcoma [6].

Conclusion

Although leiomyosarcomas are limited in the kidney, they are aggressive tumors with a poor prognosis, with a 5-year survival rate of 29-36% [1]. The majority of patients are lost within 2

years. Local recurrence is common. Distant metastases are hematogenous, and they are mostly observed in the lungs and bones.

Postoperative chemotherapy and radiotherapy can be generally performed in cases which have undergone partial resection. To date, studies published in the literature have shown that local control of the disease is better in cases of sarcoma that have undergone radical resection, but there is no survival benefit for adjuvant treatment with chemotherapy and radiotherapy. The possibility of treatment with KIT tyrosine kinase inhibitors, such as sunitinib, has been reported in phase II. Radiotherapy and chemotherapy were not applied to our case.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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